instead of penicillamine. Trientine and zinc are useful drugs for maintenance therapy. Effective treatment can reverse the neurologic features in most patients, particularly when started early. However, some patients may still progress, especially those with hepatocerebral disease. KF rings tend to decrease after 3–6 months and disappear by 2 years. Adherence to maintenance therapy is a major challenge in long-term care. Patients with advanced hepatic disease may require a liver transplant, and the potential role of organ-specific chelation therapy is under investigation.

■ NEURODEGENERATION WITH BRAIN IRON ACCUMULATION

Neurodegeneration with brain iron accumulation (NBIA) represents a group of inherited disorders characterized by iron accumulation in the basal ganglia. Clinically, they can manifest as progressive neurologic disorders with a variety of clinical features including parkinsonism, dystonia, neuropsychiatric abnormalities, and retinal degeneration. Cognitive disorders and cerebellar dysfunction may also be seen. Presentation is usually in childhood, but adult cases have been described. Multiple genes have been identified. Pantothenate kinase–associated neurodegeneration (PKAN), formerly known as Hallervorden-Spatz disease, is caused by a mutation in the *PANK2* gene, and is the most common form of NBIA, accounting for about 50% of cases. Onset is usually in early childhood and is manifest as a combination of dystonia, parkinsonism, and spasticity. MRI shows a characteristic low signal abnormality in the center of the globus pallidus on T2-weighted scans caused by iron accumulation and known as the “eye of the tiger” sign. Numerous other gene mutations have been described associated with iron accumulation including *PLA2G6, C19orf12, FA2H, ATP13A2, WDR45, FTL, CP, COASY*, and *DCAF17*. One must be cautious, however, not to assume that all cases with iron accumulation in the basal ganglia represent an NBIA, because iron accumulation in specific basal ganglia regions is normal, and excess iron accumulation may occur in the basal ganglia as a nonspecific secondary consequence of neurodegeneration unrelated to a defect in iron metabolism.